Neuromuscular Scoliosis in Children with Spinal Cord Injury

M.J. Mulcahey, PhD, OTR/L,[1,2] *John P. Gaughan, PhD,*[3] *Randal R. Betz, MD,*[2] *Amer F. Samdani, MD,*[2] *Nadia Barakat, PhD,*[2] *and Louis N. Hunter, PT, MS*[1]

[1]*Jefferson School of Health Professions, Thomas Jefferson University, Philadelphia, Pennsylvania;* [2]*Shriners Hospitals for Children, Philadelphia, Pennsylvania;* [3]*Biostatistics Consulting Center, Temple University School of Medicine, Philadelphia, Pennsylvania*

Background: The prevalence of neuromuscular scoliosis in children with spinal cord injury (SCI) is high. Published reports suggest that age at time of injury is the most important determinant. No studies have evaluated neurological characteristics using standardized methods to determine if they are strong predictors of scoliosis. **Objective:** To test the hypothesis that neurological level, motor score, and injury severity are strong predictors of neuromuscular scoliosis. **Methods:** Two hundred seventeen children were evaluated using the testing guidelines of the International Standards for Neurological Classification of Spinal Cord Injury. Cobb angles were calculated from plain radiographs as a measure of scoliosis. Multivariate analysis with statistical selection was used to determine predictors of worst Cobb angle and spinal fusion. The odds of having a spine fusion for subjects with at least 2-year follow-up and injured prior to (n=16) and after (n=91) 12 years of age were calculated. **Results:** The hypothesis was not supported. Although there was a very high prevalence (100%) of scoliosis in the study sample, age at time of injury was the only predictor of worst curve ($P < .0001$) and spine fusion ($P < .007$). The calculated odds ratio demonstrated that children injured <12 years were 3.7 times more likely to have a spine fusion (95% CI, 0.31-44.64). **Conclusion:** There is a very high prevalence of neuromuscular scoliosis in pediatric SCI. Neurological level, motor level, and severity of injury are not strong predictors. Age is the only predictor of worst curve and spine fusion. **Key words:** *neuromuscular scoliosis, outcomes, paraplegia, pediatric spinal cord injury, tetraplegia*

Neuromuscular scoliosis is a complex secondary condition of pediatric spinal cord injury (SCI)[1-4] that develops in nearly all children injured prior to skeletal maturity.[3,5,6] The secondary complications of neuromuscular scoliosis often include pelvic obliquity, skin breakdown, pulmonary compromise, and functional decline.[2] Because of the potentially devastating consequences of neuromuscular scoliosis on body functions and the high rate of complications associated with spine surgery for neuromuscular scoliosis,[7] treatment to prevent or slow the progression of neuromuscular scoliosis is a primary focus of the longitudinal care of children with SCI.

Although the evidence on the effectiveness of bracing for neuromuscular scoliosis is low,[4,8] the assumed benefits of a thoracolumbar sacral orthosis (TLSO) likely outweigh the risks. Thus, a brace is usually prescribed.[8-10] One factor thought to influence the effectiveness of bracing on neuromuscular scoliosis is curve severity; curves less than 40° have been shown to respond to brace treatment whereas curves greater than 40° have shown minimal to no response to the brace.[8] Despite any potential benefits of the brace, there are no dosing studies to inform recommendations about wearing time. As a result, recommendations for wearing time range from "while up in chair" to "23 hours a day" with many variations. Children report loss of function while wearing the TLSO, particularly function associated with wheelchair pushing, transfers in and out of the chair, and functional reach.[9] For children with hyperhidrosis or those living in warm and tropical geographical locations, tolerance to the brace can be low. Also, despite brace treatment, scoliosis in children with SCI usually progresses and requires a definitive spinal fusion to prevent progression to a magnitude that becomes life threatening.[3]

For some children with curves under 40°, alternative treatment to the brace is available in the form of surgical intervention using "growing instrumentation systems."[11] Generally, growing

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systems are designed to surgically correct the scoliosis using spinal instrumentation without performing a spinal fusion; every 3 to 6 months, the spinal instrumentation is elongated to accommodate spinal growth. Growing systems require repeated surgeries under anesthesia. Scoliosis is controlled throughout middle childhood, but a definitive spinal fusion is typically performed during middle adolescence.

Spinal fusions are effective in arresting progressive neuromuscular scoliosis, but they are not without complications. Although preoperative pulmonary status, curve angle, and age may be predictors of postoperative complications,[12,13] the risk of pulmonary complications, pseudoarthrosis, and infections is high in all children undergoing spine surgery for treatment of neuromuscular scoliosis.[7] Although rare, vision loss has also been reported as a complication of spine fusion in a child with SCI.[14]

Perhaps the greatest clinical challenge associated with managing neuromuscular scoliosis and making treatment recommendations for children with SCI is the uncertainty about which factors are most strongly associated with curve progression. Based on the literature, age at time of injury has been recognized as the strongest determinant of progression of neuromuscular scoliosis and ultimately surgical fusion.[1,2,5,6,10] However, no study has evaluated the neurological characteristics of an adequate number of children with SCI using standardized and valid methods to determine whether they are associated with progressive neuromuscular scoliosis. Over the last 5 years, we have systematically evaluated 217 children with SCI using the techniques of the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI)[15,16] and have calculated Cobb angles as measures of neuromuscular scoliosis. We hypothesized that in addition to age at injury, neurological level, motor score, and injury severity as defined by the ISNCSCI would be strong predictors of neuromuscular scoliosis and that knowledge about these SCI characteristics, as they relate to scoliosis, could inform preventative management. Thus, the purpose of this study was to investigate the determinants of neuromuscular scoliosis as a function of age at injury, sex, neurological level, motor score, and injury severity as defined by the American Spinal Injury Association Impairment Scale (AIS).[16]

Materials and Methods

Sample

This was a cross-sectional, multicenter study of subjects who were enrolled in a larger multicenter study examining the reliability of the ISNCSCI in the pediatric SCI population.[17] Full institutional review board (IRB) approval at each participating site was established. Written informed consent was obtained from all parents or legal guardians of all subjects under 18 years of age. As per IRB guidelines, written informed assent was obtained for those subjects between 7 and 18 years, and subjects older than 18 years of age provided their own written consent.

ISNCSCI

The ISNCSCI are recognized as the primary measurement and classification system of the neurological consequences of SCI.[18-24] Over the last decade, these standards have been used as a primary outcome measure for research studies and a standard assessment in the care of patients with SCI, with the most recent revisions made in 2011.[25,26] The neurological assessments of the ISNCSCI include motor, sensory, and anorectal examinations. These assessments provide the basis for classifying the neurological level (NL), motor scores (MS) and motor level (ML), sensory scores and sensory level, the zone of partial preservation, and the degree of impairment or severity of the SCI (AIS complete or incomplete).[25]

The ISNCSCI scores from a large-scale pediatric reliability study[17] were used to describe the neurological status of the subjects. All ISNCSCI exams were conducted by research and health care professionals who had completed a 2-day training session.[27] Two experienced raters confirmed the scoring and classification based on the guidelines in the 2003 ISNCSCI reference manual.[15] Information from the medical charts included sex, current age, age at injury, years since injury, and cause of injury and whether a fusion for neuromuscular scoliosis

had been performed. Coronal Cobb angles were taken directly from radiographic films. For the purpose of this study, if a patient had more than one curve, the curve with greater severity was used for analysis.

Statistical analyses

Data were transferred into SAS, version 9.1 (SAS Institute, Cary, NC). Multivariate analysis with statistical selection was used to determine predictors of worst Cobb angle and spinal fusion for the entire sample and was repeated excluding subjects with AIS D (n=21). As a way to evaluate the impact of age at injury, the analyses were repeated on a subsample of subjects who were 14 years of age or older at the time of the study and who were either injured prior to 12 years of age (n=16) or after 12 years old (n=75) with a minimum of 2-year follow-up (n=91). For both groups, the odds of having a spinal fusion for the treatment of neuromuscular scoliosis were calculated.

Results

As summarized in **Table 1**, of the 217 subjects who were included in this study, 127 (58%) were male and 90 (42%) were female, with an average age at follow-up of 13.2 years, an average age at injury of 9 years, and an average time since injury of 4.2 years. Among the subjects who were able to be classified, 105 subjects had complete injuries (AIS A) and 96 had incomplete (AIS B, C, or D), with slightly more subjects with tetraplegia (n=112) than those with paraplegia (n=95).

Of the 217 subjects, 24 underwent spinal fusion due to progressive neuromuscular scoliosis and the remaining 186 had a minimum Cobb angle of 10°, indicating that 100% of the subjects had neuromuscular scoliosis. As shown in **Table 2**, when the Cobb angle cutoff was defined as greater than 10° or if the patient had a fusion, 78% (145/186) had scoliosis. Using a cutoff angle ≥20° (value used for brace treatment) or if the patient had a fusion, 47% (87/186) of subjects had scoliosis. Last, 20% (37/186) of subjects had a curve beyond 45°, the traditional Cobb angle severity for considering spine fusion.

When grouped as an entire sample, age of injury ($P < .0001$) and AIS classification ($P < .0095$) were the only significant predictors of worse curve (**Table 3**). With the exclusion of subjects with AIS D, age at injury ($P < .0140$) was the only significant predictor of worse curve. Age at injury was also the only significant predictor of spinal fusion in the entire sample and remained as such with the exclusion of subjects with AIS D. As shown in **Table 4**, the risk of spinal fusion increased 11% for each year decrease in age at injury; this finding is consistent with or without inclusion of subjects with AIS D. Sex, motor score, and neurological level were not predictors of worse curve or spinal fusion.

To further explore the role of being injured before skeletal maturity, the cohort was limited to subjects who were past or nearing skeletal maturity (14 years old) at their follow-up appointment with a minimum of 2 years since injury (**Table 5**). Of the subjects older than 14 years of age at the time of their evaluation and injured before 12 years of age (16/43), 13% required a spine fusion compared to 4% injured after the age of 12 (27/43). The calculated odds ratio demonstrated that subjects injured before the age of 12 were 3.7 times more likely to require a spinal fusion than those injured after age 12 (95% CI, 0.31-44.64).

Discussion

The study results did not support the hypothesis that neurological level, motor scores, and severity of injury are strong predictors of neuromuscular scoliosis in children with SCI. AIS classification was a significant predictor for the entire sample, but when subjects with strong motor incomplete injuries (AIS D) were excluded, AIS classification was no longer significant. Rather, the results showed that age at injury was the only significant predictor of worst curve and of progression to spinal fusion in children with SCI. The findings also showed that children who were injured at a younger age were more likely to have progressive neuromuscular scoliosis and a definitive spinal fusion. Similar to previous reports, there was a high prevalence (100%) of scoliosis in the study sample.

Table 1. Basic demographics of patient populations

Demographics	All subjects n (%)	Fused from scoliosis n (%)	Non-fused patients n (%)
Total	217	24	186
Missing			7
Gender			
Male	127 (58)	15 (63)	110 (59)
Female	90 (42)	9 (37)	76 (41)
Missing			7
Severity			
Complete	105 (51)	10 (42)	91 (51)
Incomplete	102 (49)	14 (58)	87 (49)
Missing	10		15
AIS			
A	105 (52)	10 (42)	91 (53)
B	45 (22)	8 (33)	36 (21)
C	30 (15)	5 (21)	25 (15)
D	21 (11)	1 (4)	20 (11)
Missing	16		21
Type			
Tetra	112 (54)	11 (48)	100 (56)
Para	95 (46)	12 (52)	79 (44)
Missing	10		15
	Mean (±SD)	**Mean (±SD)**	**Mean (±SD)**
Age at follow-up	13.2 (±4.9) n=196	12.7 (±2.4) n=18	13.3 (±5.1) n=178
Age at injury	9.0 (±6.4) n=217	6.2 (±4.9) n=24	9.4 (±6.4) n=186
Years since injury	4.2 (±3.7) n=196	6.5 (±4.3) n=18	4.0 (±3.6) n=178
Motor score	44.4(±19.7) n=200	42.9 (±20.4) n=23	44.5 (±19.9) n=172
Worst curve	25.5(±22.3) n=196	66.7 (±19.3) n=18	21.3 (±17.9) n=178

Note: AIS = American Spinal Injury Association Impairment Scale; Para = paraplegia; Tetra = tetraplegia.

Table 2. Prevalence of scoliosis for Cobb angles, excluding subjects who already had spine fusion (n=24)

	All subjects	
Cobb angles	**%**	**n/N**
>10° scoliosis	78	145/186
>20° scoliosis	47	87/186
>45° scoliosis	20	37/186

Note: 10° = diagnosis of scoliosis; 20° = degree in which brace treatment usually begins; 45° = degree that reflects surgical range.

Table 3. Multiple regression with stepwise selection for predicting worse curves and by exclusion of AIS D

	Parameter estimate	Standard error	*P*	Standardized estimate
All subjects (N=176)				
Age at injury	-1.440	0.258	.0001	-0.387
AIS classification	-4.034	1.538	.0095	-0.183
Excluding AIS D (n=157)				
Age at injury	-1.518	0.275	.0140	-0.405

Note: AIS = American Spinal Injury Association Impairment Scale.

Table 4. Predictors of fusion due to scoliosis by all AIS categories and by exclusion of AIS D using multiple logistic regression with stepwise selection

Parameter	Estimate	Standard error	*P*	OR	Standardized estimate
All subjects (N=188)					
Age at injury	0.107	0.039	.007	0.899 (0.832, 0.971)	-0.363
Excluding AIS D (n=167)					
Age at injury	-0.104	0.040	.009	0.901 (0.834, 0.975)	-0.351

Note: AIS = American Spinal Injury Association Impairment Scale; OR = odds ratio.

Table 5. Odds ratio for eventual spine fusion in children with injury before or after 12 years of age who were over 14 years of age at time of evaluation with minimum of 2 years follow-up

	Fused (%)	Not fused	Total	OR	Lower 95% CI	Upper 95% CI
All AIS						
<12	2 (13)	14	16	3.72	0.31	44.64
≥12	1 (4)	26	27			
Total	3 (7)	40	43			
Excluding AIS D						
<12	1 (9)	10	11	2.50	.142	44.05
≥12	1 (4)	25	26			
Total	2 (5)	35	37			

Note: AIS = American Spinal Injury Association Impairment Scale; OR = odds ratio.

Previous research on neuromuscular scoliosis in children with SCI suggests that age at time of injury appears to be the most important determinant of progression of neuromuscular scoliosis and, ultimately, surgical fusion.[1,2,5,6,10] The results of this study, involving 217 children with SCI who had neurological evaluations by trained raters using the standard testing guidelines of the ISNCSCI, support prior studies showing, with statistical significance, that age at injury is the only predictor of neuromuscular scoliosis. This finding has important implications for clinical programs that claim rehabilitation for muscle strengthening and/or motor recovery has the potential to prevent neuromuscular scoliosis.

Research on determinants of neuromuscular scoliosis has simultaneously examined the determinants that lead to spinal fusion. This study found that the age at injury was the only determinant of spinal fusion and that children injured prior to 12 years of age were almost 4 times as likely to require spinal fusions. Although these particular results were not statistically significant, they are clinically relevant and are similar to previous studies that report a very high percentage of children injured prior to skeletal maturity will require a spinal fusion for neuromuscular scoliosis.[1,6,28] The results of this study and the collective results of other published studies underscore the importance of ongoing education for children and parents about age at injury as the strongest predictor of scoliosis and about the likelihood of it developing and requiring a spine fusion to halt progression.

There is a void in research about how to prevent or slow the progression of neuromuscular scoliosis in children with SCI. Practice patterns vary tremendously and involve, among other things, watchful waiting, the use of a TLSO, surgery, exercise, and other modalities. With such strong evidence for age as the only significant predictor of neuromuscular scoliosis and the high prevalence of neuromuscular scoliosis among children with SCI, research should focus on how best to prevent injury in young children, as they are at the highest risk. Even though the TLSO is the mainstay of treatment, there are no definitive findings to the efficacy of bracing.[1,2,8] Fusionless surgical techniques have been described as an alternative to bracing, but they have not been adequately studied with respect to risk-benefit or effectiveness in preventing progression and spinal fusion.

There are several limitations of this study. First, the number of subjects in the subanalyses was small. There were only 21 subjects (11%) with AIS classification D. For the analysis involving a minimum of 2 years follow-up, there were only 16 subjects in the group injured who were younger than 12 years and only 27 who were injured older than 12 years. Research should examine the relationship of motor activity and ambulatory status to neuromuscular scoliosis, particularly since AIS classification was a predictor when the entire sample was considered. Another notable limitation of the study is the lack of information on the functional abilities of the subjects with incomplete injuries, specifically their ambulatory status, which is thought to have a mitigating effect on neuromuscular scoliosis. Future work should include a large number of children with incomplete injuries and a prospective study to follow the development of scoliosis and to document treatment from the time of the initial injury. Finally, the subjects in this study participate in various treatments to slow the progression of the scoliosis; these treatments were not quantified nor were they evaluated with respect to follow-through and adherence.

Conclusion

Neuromuscular scoliosis is highly prevalent among persons with SCI who are injured at a young age. Neurological level, motor score, and severity of injury are not predictors of neuromuscular scoliosis, thus altering them will not likely change the outcomes with respect to neuromuscular scoliosis. Age at injury is the only predictor of worst curve and spinal fusion in children with SCI. There is a need for prospective studies on how to prevent neuromuscular scoliosis in young children with SCI, because they are at greatest risk for developing it and ultimately requiring surgical spine fusion.

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REFERENCES

1. Dearolf WW 3rd, Betz RR, Vogel LC, Levin J, Clancy M, Steel HH. Scoliosis in pediatric spinal cord-injured patients. *J Pediatr Orthop.* 1990;10(2):214-218.
2. Driscoll SW, Skinner J. Musculoskeletal complications of neuromuscular disease in children. *Phys Med Rehabil Clin N Am.* 2008;19(1):163-194.
3. Parent S, Dimer J, Dekutoski M, Roy-Beaudry M. Unique feature of pediatric spinal cord injury. *Spine.* 2010;35(21 suppl):S202-208.
4. Parent S, Mac-Thiong JM, Roy-Beaudry M, Sosa JF, Labelle H. Spinal cord injury in the pediatric population: a systematic review of the literature. *J Neurotrauma.* 2011;28(8):1515-1524.
5. Lancourt JE, Dickson JH, Carter RE. Paralytic spinal deformity following traumatic spinal cord injury in children and adolescents. *J Bone Joint Surg Am.* 1981;63(1):47-53.
6. Mayfield JK, Erkkila JC, Winter RB. Spine deformity subsequent to acquired childhood spinal cord injury. *J Bone Joint Surg Am.* 1981;63(9):1401-1411.
7. Sharma S, Wu C, Anderson T, Wang Y, Hansen ES, Bunger CE. Prevalence of complications in neuromuscular scoliosis surgery: a meta-analysis from the past 15 years [published online ahead of print October 21, 2012]. *Eur Spine J.*
8. Mehta S, Betz RR, Mulcahey MJ, McDonald C, Vogel LC, Anderson C. Effect of bracing on paralytic scoliosis secondary to spinal cord injury. *J Spinal Cord Med.* 2004;27(suppl 1):S88-92.
9. Chafetz R, Mulcahey MJ, Betz RR, et al. Impact of prophylactic thoracolumbosacral bracing on function and activities of daily living in the pediatric spinal cord injured population. *J Spinal Cord Med.* 2007;(suppl 1):S178-S183.
10. Tsirikos AI, Markham P, McMaster MJ. Surgical correction of spinal deformities following spinal cord injury occurring in childhood. *J Surg Orthop Adv.* 2007;16(4):174-186.
11. Samdani AF, Ranade A, Dolch HJ, et al. Bilateral use of vertical expandable prosthetic titanium rib attached to the pelvis: a novel treatment for scoliosis in the growing spine. *J Neurosurg Spine.* 2009;10(4):287-292.
12. Kang GR, Suh SW, Lee OI. Preoperative predictors of postoperative pulmonary complications in neuromuscular scoliosis. *J Orthop Sci.* 2011;16(2):139-147.
13. Master DL, Son-Hing JP, Poe-Kochert C, Armstrong DG, Thompson GH. Risk factors for major complications for neuromuscular scoliosis. *Spine.* 2011;36(7):564-571.
14. Samdani AF, Rutter L, Betz RR, Mulcahey MJ. Vision loss after spine fusion for scoliosis in a child with spinal cord injury. *J Spinal Cord Med.* 2009;32(5):591-594.
15. American Spinal Injury Association. *Reference Manual for the International Standards for Neurological Classification of Spinal Cord Injury.* Chicago: Author; 2003.
16. Marino RJ, Barros T, Biering-Sorensen F, et al. International Standards for Neurological Classification of Spinal Cord Injury. *J Spinal Cord Med.* 2003;26(suppl 1):S50-56.
17. Mulcahey MJ, Vogel L, Betz R, Samdani A, Chafetz R, Gaughan J. The International Standards for Neurological Classification of Spinal Cord Injury: psychometric evaluation and guidelines for use with children and youth. *Phys Med Rehabil.* 2011; 92:1264-1269.
18. Bracken MB, Shepard MJ, Holford TR, et al. Administration of methylprednisolone for 24 or 48 hours or tirilazad mesylate for 48 hours in the treatment of acute spinal cord injury. Results of the third national acute spinal cord injury randomized controlled trial. National Acute Spinal Cord Injury Study. *JAMA.* 1997;277(20):1597-1604.
19. Maynard FM Jr, Bracken MB, Creasey G, et al. International Standards for Neurological and Functional Classification of Spinal Cord Injury. American Spinal Injury Association. *Spinal Cord.* 1997;35(5):266-274.
20. Bracken MB, Shepard MJ, Collins WF, et al. A randomized, controlled trial of methylprednisolone or naloxone in the treatment of acute spinal-cord injury. Results of the second National Acute Spinal Cord Injury Study. *N Engl J Med.* 1990;322(20):1405-1411.
21. Baptiste DC, Fehlings MG. Pharmacological approaches to repair the injured spinal cord. *J Neurotrauma.* 2006;23(3-4):318-334.
22. Brown PJ, Marino RJ, Herbison GJ, Ditunno JF Jr. The 72-hour examination as a predictor of recovery in motor complete quadriplegia. *Arch Phys Med Rehabil.* 1991;72(8):546-548.
23. Blaustein DM, Zafonte R, Thomas D, Herbison GJ, Ditunno JF. Predicting recovery of motor complete quadriplegic patients. 24 hour v 72 hour motor index scores. *Am J Phys Med Rehabil.* 1993;72(5):306-311.
24. McDonald JW, Becker D, Sadowsky CL, Jane JA Sr, Conturo TE, Schultz LM. Late recovery following spinal cord injury. Case report and review of the literature. *J Neurosurg.* 2002;97(2 suppl):252-265.

25. American Spinal Injury Association. *Reference Manual for the International Standards for Neurological Classification of Spinal Cord Injury*. Atlanta: Author; 2011.
26. Kirshblum SC, Waring W, Biering-Sorensen F, et al. Reference for the 2011 revision of the International Standards for Neurological Classification of Spinal Cord Injury. *J Spinal Cord Med*. 2011;34(6):547-554.
27. Chafetz R, Vogel L, Betz R, Gaughan J, Mulcahey M. International Standards for Neurological Classification of Spinal Cord Injury: training effect on accurate classification. *J Spinal Cord Med*. 2008;31(5):538–542.
28. Campbell J, Bonnett C. Spinal cord injury in children. *Clin Orthop Rel Res*. 1975;(112):114-123.